Case Report

Postmortem diagnosis of massive gastrointestinal bleeding in a patient with aberrant right subclavian artery–esophageal fistula

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Case: Aberrant right subclavian artery–esophageal fistula is a rare, but fatal, complication. A 55-year-old febrile man with a nasogastric feeding tube developed sudden massive hematemesis and shock.

Outcome: Upper endoscopy revealed an intragastric hematoma with no active bleeding observed except for oozing from an esophageal tear. Enhanced computed tomography scan detected aberrant right subclavian artery but was unable to determine the bleeding source. Repeat endoscopy carried out on day 2 confirmed hemostasis and the disappearance of the intragastric hematoma. However, the patient suddenly developed recurrent massive hematemesis with refractory shock on day 4 and died. Postmortem computed tomography revealed endoscopic clips in contiguity with aberrant right subclavian artery; a final diagnosis of aberrant right subclavian artery–esophageal fistula was made.

Conclusion: Our case demonstrates aberrant right subclavian artery–esophageal fistula may present with transient spontaneous hematemesis in a state of shock, possibly related to fever of unknown origin, and is challenging to diagnose by repeated endoscopy once hematemesis develops.

Key words: Artery–esophageal fistula, computed tomography, nasogastric tube, unexplained gastrointestinal bleeding, upper endoscopy

INTRODUCTION

ABERRANT RIGHT SUBCLAVIAN artery (ARSA) is the most common abnormality of the aortic arch and has a 0.5–1.8% prevalence in the general population.[1] In a majority of cases, the location is posterior to the esophagus, making ARSA particularly susceptible to extrinsic compression and pressure necrosis secondary to placement of devices, such as nasogastric (NG) tubes;[2] thus, predisposing towards the formation of ARSA–esophageal fistula. Upper gastrointestinal (GI) bleeding in ARSA–esophageal fistula is an exceedingly rare occurrence, but its mortality rate reaches almost 100% as bleeding is usually massive and sudden.[3] This paper describes the postmortem diagnosis of a case of this devastating complication diagnosed postmortem in a 55-year-old man.

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CASE

A 55-YEAR-OLD MAN, BEDRIDDEN for the previous 3 months due to brain hemorrhage, developed sudden massive hematemesis and was brought to our hospital. He was tracheostomized and received enteral alimentation by NG tube. He had unexplained intermittent fever over the preceding week for which he received empiric antimicrobial without culture studies. On admission, he was tachypneic with a respiratory rate of 24 breaths/min. His blood pressure was 114/65 mmHg with a heart rate of 114 b.p.m. The consciousness level was E3VTM4 on the Glasgow Coma Scale, usual for this patient, and the body temperature was 36.4°C. Laboratory examinations revealed the following: white blood cell count, 12,620/μL; hemoglobin, 6.6 g/dL; platelets, 18.1×10^4/μL; blood urea nitrogen, 26 mg/dL; creatinine, 0.66 mg/dL; and C-reactive protein, 0.55 mg/dL. Coagulation studies were normal.

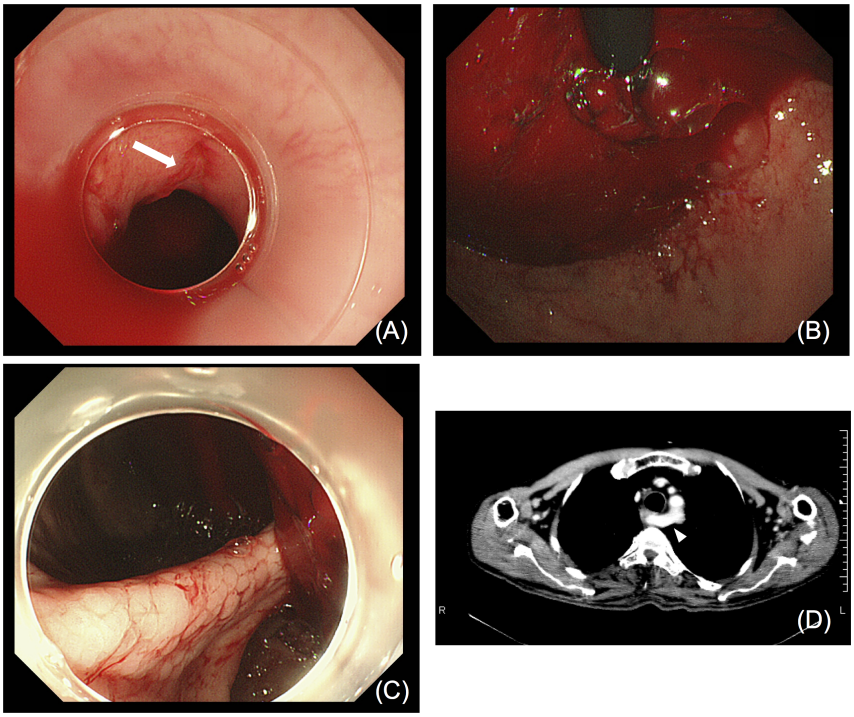

Fig. 1. Upper endoscopy on admission of a 55-year-old febrile man with a nasogastric feeding tube (A–C) revealed upper gastrointestinal bleeding; however, the bleeding site remained unidentified as the examination was insufficient due to massive hematoma. An esophageal tear was also found (arrow). Contrast-enhanced computed tomography after endoscopy (D) did not visualize extravasation; however, an aberrant right subclavian artery was revealed (arrowhead).

At the emergency room, he had recurrent massive bloody emesis with severe shock (blood pressure, 51/20 mmHg; heart rate, 68 b.p.m.) and was resuscitated. He underwent urgent upper endoscopy with over-tube from the start that revealed upper GI bleeding (Fig. 1A–C); however, the bleeding source was not identified due to a huge hematoma in the stomach. Esophageal tear with oozing blood was also shown, however, the site of active bleeding was not identified and endoscopic hemostasis was not carried out. Contrast-enhanced computed tomography (CT) showed no extravasation. An ARSA in paper-thin contact with the esophagus was revealed (Fig. 1D), however, we were unable to ascertain its relationship with the GI bleeding as no contrast medium was visualized in the intestinal lumen. The patient was stabilized after receiving a total of 10 U packed red blood cells and 6 U fresh frozen plasma and treatment with a proton pump inhibitor was initiated.

His hemodynamic status was stable for over 36 h following admission to the intensive care unit. Follow-up upper endoscopy carried out on day 2 did not show active bleeding, however, a scar in the middle of the esophagus was observed and thought to possibly be due to contact injury from the over-tube procedure (Fig. 2A–F). Despite receiving continuous proton pump inhibitor treatment, anemia progressed on day 3 (hemoglobin, 8.4–6.0 g/dL) without hemodynamic instability, for which he received an additional transfusion of 2 U packed red blood cells. However, he had a second episode of massive hematemesis and went into shock (blood pressure, 60/38 mmHg; heart rate, 111 b.p.m.) on day 4. Urgent upper endoscopy was performed for the third time indicating the small laceration, seen during the preceding endoscopy, had been exacerbated with massive blowout bleeding in the middle of the esophagus. This lesion was obviously distinct from the known esophageal tear (Fig. 3A,B). Endoscopic hemostasis was unsuccessful. He went into refractory shock and instantly into cardiac arrest. His hemodynamic instability precluded surgical intervention or interventional radiology procedure; eventually, the patient died. Autopsy CT scans revealed an endoscopic clip in contiguity with the ARSA (Fig. 3C). Additionally, re-evaluation of a reconstructed CT of the ARSA on admission showed a bleb in contact with the esophagus; a diagnosis of artery esophageal fistula secondary to an ARSA was made postmortem (Fig. 3D).

DISCUSSION

WE REPORT A postmortem case of ARSA–esophageal fistula that provided three important clinical insights: (i) GI bleeding from ARSA–esophageal fistula can present as spontaneous hematemesis with shock, (ii) once hemostasis is achieved, it is difficult to diagnose by endoscopy, (iii) fever of unknown origin may be a predictor of ARSA–esophageal fistula in patients with prolonged NG tube insertion.

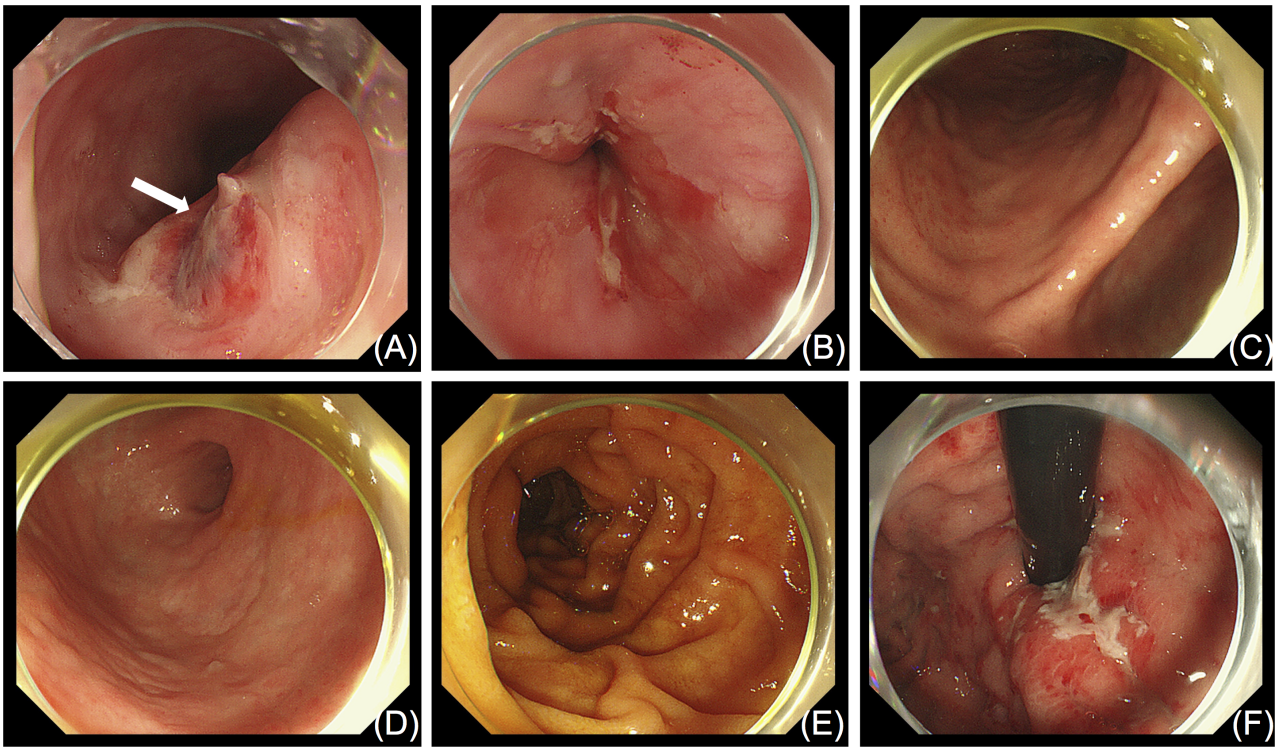

Fig. 2. Follow-up endoscopy on hospital day 2 of a 55-year-old febrile man with a nasogastric feeding tube (A–F) did not show active bleeding and the huge hematoma shown on admission disappeared. Also, a healing esophageal tear (B) and a possible over-tube injury appearing as a small protrusion due to mucosal detachment following abrasion by the over-tube (arrowhead) were seen without evidence of bleeding.

In aortoesophageal fistula, sentinel bleeding may occur in up to 63% of cases; however, few cases of ARSA–esophageal fistula involve clear sentinel bleeding.[2,4] Hematemesis is usually sudden and massive, therefore patients commonly go into instantaneous refractory shock.[2,3] Urgent endoscopy may be used to rule out other causes of GI bleeding and allow localization and control of bleeding sites using balloon compression, although hemostatic control from the GI side was almost certainly impossible in this case.[1–3] This case suggests the possibility of spontaneous hematemesis from an ARSA–esophageal fistula with severe shock (near cardiac arrest), albeit over a limited period.

The present case also demonstrated that use of an over-tube may lead to signs of esophageal fistula being overlooked, possibly leading to misdiagnosis of ARSA–esophageal fistula as an over-tube injury on follow-up endoscopy. Previous studies have reported the diagnostic sensitivity of endoscopy for aortoesophageal fistula to be only 38%.[1,5] Aberrant right subclavian artery should be included in the differential diagnoses when investigating upper GI bleeding in patients with relevant risk factors and hemodynamic instability.[1,3] In retrospect, the right side of the esophageal wall (shown in Fig. 2A) was elevated with an appearance in keeping with a submucosal tumor. The erythematous region indicates the location of the ARSA under the esophageal mucosa. The central, erythematous, irregular elevation also indicated the region of exposed vasculature in the ARSR–esophageal fistula. Furthermore, the contact injury of NG tube insertion was one possible cause of the mucosal fragility followed by ARSA fistula. Although plain chest CT can easily identify ARSA, it is often not feasible during emergency cases, particularly in unstable patients. Regardless, fistula between an ARSA and the esophagus should be suspected when there is massive, bright-red hematemesis.[6] Furthermore, even where hemostasis is achieved, the presence of massive hematoma, which may mask endoscopic findings, should trigger suspicion of an aortoesophageal fistula.

Finally, aortogastrointestinal tract fistula has been shown to contribute to intravascular infection[7] and may manifest as fever of unknown origin with ready initiation of antimicrobials. In this case, the patient had unexplained intermittent

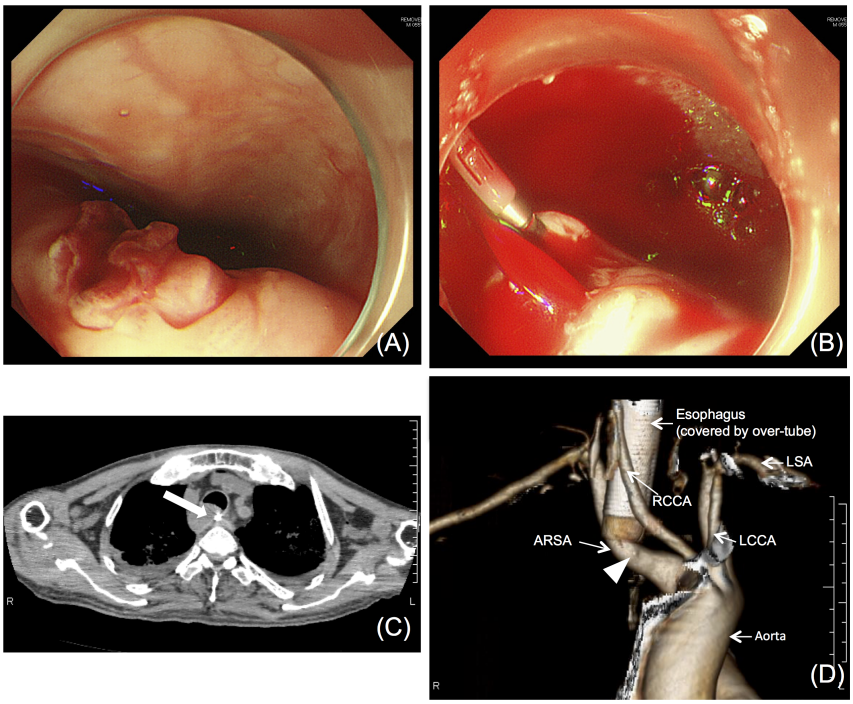

Fig. 3. Third endoscopy of a 55-year-old febrile man with a nasogastric feeding tube (A, B) revealed massive blowout bleeding from the lesion was initially considered an over-tube injury during previous examinations. Post-mortem computed tomography carried out on autopsy (C) revealed an endoscopic clip located adjacent to the aberrant right subclavian artery (ARSA) (arrow). An arterial bleb (arrowhead) in contact with the esophagus was revealed by reconstruction of computed tomography images on admission (D). LCCA, light common carotid artery; LSA, left subclavian artery; RCCA, right common carotid artery.

fever for over 1 week prior to admission and received empiric antimicrobial therapy. There was evidence of bacterial infection in this case, implying that clinicians should contemplate the possibility of an aortoesophageal fistula in febrile patients with ARSA.

CONCLUSION

WE SHOWED THAT ARSA–esophageal fistula can lead to transient spontaneous hematemesis with shock. Gastrostomy may be a better choice than long-term tube placement when ARSA in paper-thin contact with the esophagus is found. Appropriate management of these patients requires a high index of suspicion in the setting of massive arterial hematemesis. Hemostasis with a multidisciplinary approach consisting of either endoscopic balloon compression with angiographic control or urgent surgery, or both, is recommended.[1,3]

CONFLICT OF INTEREST

NONE.

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